Complete Summary

GUIDELINE TITLE

Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia: an evidence-based review.

BIBLIOGRAPHIC SOURCE(S)

RF/RHD Guideline Development Working Group of the National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand. Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia: an evidence-based review. Sydney (Australia): National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand; 2006 Jun. 84 p. [261 references]

GUIDELINE STATUS

This is the current release of the guideline.

** REGULATORY ALERT **

FDA WARNING/REGULATORY ALERT

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- February 28, 2008, Heparin Sodium Injection: The U.S. Food and Drug Administration (FDA) informed the public that Baxter Healthcare Corporation has voluntarily recalled all of their multi-dose and single-use vials of heparin sodium for injection and their heparin lock flush solutions. Alternate heparin manufacturers are expected to be able to increase heparin production sufficiently to supply the U.S. market. There have been reports of serious adverse events including allergic or hypersensitivity-type reactions, with symptoms of oral swelling, nausea, vomiting, sweating, shortness of breath, and cases of severe hypotension.
- August 16, 2007, Coumadin (Warfarin): Updates to the labeling for Coumadin to include pharmacogenomics information to explain that people's genetic makeup may influence how they respond to the drug.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

- Acute rheumatic fever
- Rheumatic heart disease

GUIDELINE CATEGORY

Diagnosis
Management
Prevention
Treatment

CLINICAL SPECIALTY

Cardiology
Infectious Diseases
Internal Medicine
Pediatrics
Rheumatology

INTENDED USERS

Advanced Practice Nurses
Allied Health Personnel
Health Care Providers
Nurses
Physician Assistants
Physicians

GUIDELINE OBJECTIVE(S)

- To identify the standard of care, including preventive care, that should be available to all people
- To identify areas where current management strategies may not be in line with available evidence
- To ensure, in the interests of equity, that high-risk populations receive the same standard of care as that available to other Australians

TARGET POPULATION

Patients with acute rheumatic fever and rheumatic heart disease

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis of Acute Rheumatic Fever (ARF)

1. Diagnosis using the Jones criteria and World Health Organization (WHO)
2. Differential diagnosis
3. Echocardiography

Management of ARF

1. Confirm diagnosis
2. Non-steroidal anti-inflammatory drugs, paracetamol, and corticosteroids
3. Hospitalization

Secondary Prevention and Rheumatic Heart Disease (RHD) Control

1. Benzathine penicillin G (BPG)
2. Alternatives to BPG for special populations, including oral penicillin in patients who refuse BPG, referral to allergist for patients allergic to penicillin, and non-beta-lactam antimicrobial for patients with confirmed reaction to penicillin
3. Patient education and promotion of continuing adherence
4. Implementation of a coordinated control program, including specialist review and echocardiography

Diagnosis and Management of chronic RHD

General

1. Access to a specialist and/or cardiologist
2. Access to echocardiography
3. Monitoring of anticoagulation therapy
4. Secondary prophylaxis with penicillin

Mitral Regurgitation

1. Echocardiography
2. Diuretic therapy and angiotensin-converting enzyme (ACE) inhibitors
3. Referral for surgery (mitral valve repair or replacement)

Mitral Stenosis

1. Doppler and two-dimensional echocardiography
2. Interventional therapy including diuretic therapy in patients who develop congestive heart failure
3. Warfarin to treat atrial fibrillation
4. Direct-current cardioversion
5. Percutaneous balloon mitral valvuloplasty
6. Valve repair

Aortic Regurgitation

1. Echocardiography
2. Vasodilator therapy
3. Nifedipine or ACE inhibitors
4. Referral for surgery (aortic valve replacement, repair, or Ross procedure)

Aortic Stenosis

1. Two-dimensional and continuous wave echocardiography
2. Percutaneous aortic valvuloplasty
3. Aortic valve replacement

Pregnancy and RHD

1. Percutaneous balloon mitral valvuloplasty in patients with moderate or severe mitral stenosis
2. Low molecular weight heparin, warfarin or low molecular weight heparin then warfarin

MAJOR OUTCOMES CONSIDERED

- Sensitivity and specificity of diagnostic tests
- Adherence to secondary prophylaxis
- Rates of further attacks of acute rheumatic fever (ARF), cardiac damage, and premature death
- Rates of surgery

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

I: Evidence obtained from a systematic review of all relevant randomized controlled trials (RCTs)

II: Evidence obtained from at least one properly designed randomised controlled trial

III-1: Evidence obtained from well-designed pseudo-randomised controlled trials (alternate allocation or some other method)

III-2: Evidence obtained from comparative studies with concurrent controls and allocation not randomised (cohort studies), case-control studies, or interrupted time series without a control group

III-3: Evidence obtained from comparative studies with historical control, two or more single-arm studies, or interrupted time series without a parallel control group

IV: Evidence obtained from case series, either post-test or pre-test and post-test

METHODS USED TO ANALYZE THE EVIDENCE

Review of Published Meta-Analyses
Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

This review was jointly developed by the National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. The review is the product of a rigorous and iterative process:

- A writing group comprising experts in the area prepared an initial draft review.
- Selected individuals with experience in acute rheumatic fever/rheumatic heart disease (ARF/RHD) management then reviewed each chapter, and their suggestions were incorporated into a second draft.
- The revised draft was widely distributed to a range of stakeholders, who were then invited to a one-day workshop in November 2004.
- The stakeholders reviewed the draft and reached consensus on areas of disagreement.
- A third draft was then prepared and re-distributed for further comment.
- Comments were then incorporated into a final draft, which was endorsed by the stakeholders.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Grades of Recommendations

A. Rich body of high-quality randomized controlled trial (RCT) data
B. Limited body of RCT data or high-quality non-RCT data
C. Limited evidence
D. No evidence available—panel consensus judgment

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The revised draft was widely distributed to a range of stakeholders, who were then invited to a one-day workshop in November 2004.The stakeholders reviewed the draft and reached consensus on areas of disagreement. A third draft was then prepared and re-distributed for further comment. Comments were then incorporated into a final draft, which was endorsed by the stakeholders.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Note from the National Guideline Clearinghouse: The recommendations that follow are from the guideline's "Summary of Key Recommendations"; detailed graded recommendations can be found in the original guideline document.

Diagnosis and Management of Acute Rheumatic Fever

Acute rheumatic fever (ARF) is an auto-immune response to bacterial infection with group A streptococcus (GAS). People with ARF are often in great pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF leaves no lasting damage to the brain, joints, or skin. However, rheumatic heart disease (RHD) may persist. People who have had ARF previously are much more likely than the wider community to have subsequent episodes. Recurrences of ARF may cause further valve damage, leading to steady worsening of RHD.

Although the exact causal pathway is unknown, it seems that some strains of GAS are "rheumatogenic" and that a small proportion of people in any population (3–5%) have an inherent susceptibility to ARF.

While it is widely thought that only upper respiratory tract infection with GAS can cause ARF, there is evidence that GAS skin infections may play a role in certain populations, including Aboriginal and Torres Strait Islander Australians.

ARF is predominantly a disease of children aged 5–14 years, although people can have recurrent episodes well into their forties. The prevalence of RHD peaks in the third and fourth decades. Therefore, although ARF is a disease with its roots in childhood, its effects are felt throughout adulthood, especially in the young adult years when people might otherwise be at their most productive.

Diagnosis of ARF

Accurate diagnosis of ARF is important. Over-diagnosis results in unnecessary treatment over a long time, while under-diagnosis leads to further attacks of ARF, cardiac damage, and premature death. Diagnosis remains a clinical decision, as there is no specific laboratory test.

The diagnosis of ARF is usually guided by the Jones criteria and the more recent World Health Organization (WHO) criteria. In this guideline, the Jones and WHO criteria have been further modified to form the 2006 Australian criteria for the diagnosis of acute rheumatic fever.

Many medical practitioners in Australia have never seen a case of ARF, because the disease has largely disappeared from the populations among which they train and work. It is very important that health staff receive appropriate education about ARF before postings to remote areas.

Many of the clinical features of ARF are non-specific, so a wide range of differential diagnoses should be considered. In a region with high compared to low incidence of ARF, a person with fever and arthritis is more likely to have ARF. Some post-streptococcal syndromes may be confused with ARF but these diagnoses should rarely, if ever, be made in high-risk populations.

All patients with suspected or confirmed ARF should undergo echocardiography, if available, to confirm or refute the diagnosis of rheumatic carditis. Echocardiographic evidence of valve damage (subclinical or otherwise), diagnosed by a clinician with experience in ARF and RHD, may be included as a major manifestation in the diagnosis of ARF.

Management of ARF

In the first few days after presentation, the major priority is confirming the diagnosis. With the exception of heart failure management, none of the treatments offered to patients with ARF has been proven to alter the outcome of the acute episode or the amount of damage to heart valves. Thus, there is no urgency to begin definitive treatment. Non-steroidal anti-inflammatory drugs reduce the pain of arthritis, arthralgia, and fever of ARF, but can confuse the diagnosis. Paracetamol and codeine are recommended for pain relief until the diagnosis is confirmed. Corticosteroids are sometimes used for severe carditis, although there is no evidence that they alter the longer-term outcome.

Ideally, all patients with suspected ARF (first episode or recurrence) should be hospitalized as soon as possible after onset of symptoms. This ensures that all investigations are performed and, if necessary, the patient observed to confirm the diagnosis before commencing treatment.

Secondary Prevention and Rheumatic Heart Disease Control

Secondary prevention refers to the early detection of disease and implementation of measures to prevent recurrent and worsening disease.

Secondary prophylaxis with benzathine penicillin G (BPG) is the only RHD control strategy shown to be effective and cost-effective at both community and population levels. Randomised controlled trials have shown that regular administration is required to prevent recurrent ARF.

Secondary Prophylaxis

Secondary prophylaxis with BPG is recommended for all people with a history of ARF or RHD. Four-weekly BPG is currently the treatment of choice, except in patients considered to be at high risk, for whom 3-weekly administration is recommended. The benefits of 3-weekly BPG injections are offset by the difficulties of achieving good adherence, even to the standard 4-weekly regimen. Prospective data from New Zealand showed that few, if any, recurrences occurred among people who fully adhered to a 4-weekly BPG regimen.

Alternatives to BPG are available, although they are less effective and require careful monitoring.

- *In patients who refuse intramuscular BPG*, oral penicillin can be offered, although it is less effective than BPG in preventing GAS infections and subsequent recurrences of ARF. For patients taking oral penicillin, the consequences of missed doses must be emphasised, and adherence monitored.
- *In patients who may be allergic to penicillin*, an allergist should be consulted. The rates of allergic and anaphylactic reactions to monthly BPG are low, and fatal reactions are exceptionally rare. There is no increased risk with prolonged BPG use.
- *In patients with a confirmed, immediate, and severe allergic reaction to penicillin*, a nonbeta-lactam antimicrobial (e.g., erythromycin) should be used instead of BPG.
- *In pregnant patients*, penicillin prophylaxis should continue for the duration of pregnancy to prevent recurrent ARF. There is no evidence of teratogenicity. Erythromycin is also considered safe in pregnancy, although controlled trials have not been conducted.
- *In anticoagulated patients*, BPG injections should be continued unless there is evidence of uncontrolled bleeding, or the international normalised ratio is outside the defined therapeutic window. Intramuscular bleeding is rare when BPG injections are used in conjunction with anti-coagulation therapy.

The appropriate duration of secondary prophylaxis is determined by age, time since the last episode of ARF, and potential harm from recurrent ARF.

All people with ARF or RHD should continue secondary prophylaxis for a minimum of 10 years after the last episode of ARF or until the age of 21 years (whichever is longer). Those with moderate or severe RHD should continue secondary prophylaxis up to the age of 35–40 years.

Infective endocarditis is a dangerous complication of RHD and a common adverse event following prosthetic valve replacement in Aboriginal and Torres Strait Islander Australians. People with established RHD or prosthetic valves should receive antibiotic prophylaxis prior to procedures expected to produce bacteraemia (e.g., dental procedures, surgical procedures where infection is present).

Adherence to Secondary Prophylaxis

Persistent high rates of recurrent ARF in Australia highlight the continued failure of secondary prevention. In the Top End of the Northern Territory in the 1990s, 28% of patients on secondary prophylaxis missed half or more of their scheduled BPG injections over a 12-month period, while 45% of all episodes of ARF were recurrences.

A variety of factors, mainly sociological, combine to limit the effectiveness of secondary prophylaxis. The major reasons for poor adherence in remote Australian Aboriginal and Torres Strait Islander communities are the availability and acceptability of health services, rather than personal factors such as injection refusal, pain of injections, or a lack of knowledge or understanding of ARF and RHD. Adherence is improved when patients feel a sense of personalised care and "belonging" to the clinic, and when recall systems extend beyond the boundaries of the community.

Hospitalisation for ARF provides an ideal opportunity to begin secondary prophylaxis, and to educate patients and families on how important it is to prevent future episodes of ARF. Continuing education and support by primary care staff, using culturally appropriate educational materials, should follow once the patient has returned home.

Secondary prevention of further episodes of ARF is a priority. It should include strategies aimed at improving the delivery of secondary prophylaxis and patient care, the provision of education, coordinating available health services, and advocacy for necessary and appropriate resources.

Strategies to promote continuing adherence include:

- Routine review and care planning
- Recall and reminder systems
- Having local staff members dedicated to secondary prophylaxis and coordinating routine care
- Supporting and utilising the expertise, experience, community knowledge, and language skills of Aboriginal health workers
- Improving staff awareness of diagnosis and management of ARF and RHD
- Taking measures to minimise staff turnover

- Implementing measures to reduce the pain of injections (e.g., use a 23-gauge needle, warm syringe to room temperature, apply pressure with thumb before inserting needle, deliver injection very slowly)

RHD Control Programs

A coordinated control program, including specialist review and echocardiography, is the most effective approach to improving BPG adherence and clinical follow-up of people with RHD.

Recommended elements of RHD control programs include the following:

- A single, centralised (preferably computerised) ARF/RHD register for each program
- A dedicated coordinator (this is critical to the success of the program)
- Integration of activities into the established health system to ensure the control program continues to function well despite staffing changes

Control programs for ARF and RHD should be evaluated using criteria for routine care and key epidemiological objectives.

Diagnosis and Management of Chronic Rheumatic Heart Disease

It is difficult and expensive for Aboriginal and Torres Strait Islander people to travel to major centres for cardiac services, which are often hospital based. Although specialist outreach services are improving in many regions, access to specialist care is suboptimal in rural and remote areas.

Implementing guidelines on the diagnosis and management of chronic RHD has major implications for Aboriginal and Torres Strait Islander health care services, especially in rural and remote regions. In addition to access to appropriate primary care services, best practice for RHD requires:

- Access to a specialist physician and/or cardiologist (preferably the same specialist over a long time)
- Access to echocardiography—portable echocardiography may be required so that all RHD patients in Australia have access to echocardiography, regardless of location
- Adequate monitoring of anticoagulation therapy in patients with atrial fibrillation and/or mechanical prosthetic valves
- Secondary prevention with penicillin prophylaxis

All patients with murmurs suggestive of valve disease, or a past history of rheumatic fever, require echocardiography. This will detect any valvular lesion, and allow assessment of its severity and of left ventricular (LV) size and systolic function. Serial echocardiographic data play a critical role in helping to determine the timing of surgical intervention.

The fundamental goal in long-term management of chronic RHD is to avoid, or at least delay, valve surgery. Therefore, prophylaxis with BPG to prevent recurrent ARF is a crucial strategy in managing patients with chronic RHD. Where adherence

to secondary prevention is poor, there is greater need for surgical intervention, and long-term surgical outcomes are not as good.

Valvular Lesions in RHD

Mitral Regurgitation

Mitral regurgitation is the most common valvular lesion in RHD, particularly in young patients. In chronic mitral regurgitation, volume overload of the left ventricle and left atrium occurs, which in more severe cases eventually results in a progressive decline in systolic contractile function. Patients with mild or moderate mitral regurgitation may remain asymptomatic for many years. Initial symptoms include dyspnoea on exertion, fatigue, and weakness, and these may progress slowly over time or worsen after a recurrence of rheumatic fever, chordal rupture, or onset of atrial fibrillation.

There is wide individual variation in the rate of progression of mitral regurgitation, although many cases tend to progress over 5–10 years, especially if there is a recurrence of ARF.

Key points in diagnosis and management of mitral regurgitation include the following.

- Echocardiography is used to confirm the diagnosis, quantify the severity of regurgitation, and assess LV size and function. In asymptomatic and mildly symptomatic patients with moderate or more severe mitral regurgitation, echocardiography should be performed at least every 6–12 months.
- Clinical heart failure requires diuretic therapy and angiotensin-converting enzyme (ACE) inhibitors.
- Patients with severe mitral regurgitation should be referred for surgery if they become symptomatic or if they have echocardiographic indicators of reduced LV systolic function or an end systolic diameter by echo of $\geq$40 mm. Patients who are asymptomatic or mildly symptomatic and have severe mitral regurgitation and normal LV systolic function should consult cardiac surgeons early, so that appropriate care plans can be developed.
- Mitral valve repair rather than replacement is the operation of choice for symptomatic dominant or pure mitral regurgitation. If the mitral valve is not suitable for repair, the options are valve replacement, either with a mechanical valve prosthesis or a bioprosthetic valve.

Mitral Stenosis

In mitral stenosis, progressive obstruction to LV inflow develops due to fibrosis and partial fusion of the mitral valve leaflets. Approximately 30% of Aboriginal RHD patients in the Northern Territory aged 10–19 years have mitral stenosis, and the mean age of those with mitral stenosis is 33 years. In the Aboriginal and Torres Strait Islander population, mitral stenosis progresses more rapidly than in the non-Aboriginal and Torres Strait Islander population and patients become symptomatic at a younger age. More rapid progression may be due to undetected recurrences of rheumatic fever.

The initial symptom is exertional dyspnoea, which worsens slowly over time. Symptoms of heart failure (orthopnoea, paroxysmal dyspnoea and occasionally haemoptysis) develop as the mitral valve orifice decreases to less than 1.0–1.5 cm.

Key points in diagnosis and management of mitral stenosis include the following.

- Doppler and two-dimensional echocardiography is used to quantitate the severity of mitral stenosis; assess associated valve lesions, LV function, left atrial size; and estimate pulmonary artery systolic pressure.
- The treatment of symptomatic moderate to severe mitral stenosis is interventional therapy. Patients who develop congestive heart failure respond to diuretic therapy.
- Atrial fibrillation is the most common complication of mitral stenosis, requiring long-term prophylactic anticoagulation with warfarin. When new-onset atrial fibrillation is associated with symptoms, consideration should be given to direct-current cardioversion to restore sinus rhythm.
- Percutaneous balloon mitral valvuloplasty is the treatment of choice for dominant or pure mitral stenosis. The indication is a mitral valve area <1.5 cm^2 with progressive symptoms, or if asymptomatic, a history of thromboembolism or significant pulmonary hypertension.
- The short-term and medium-term results are comparable to surgical valvuloplasty, with 65% of patients being free of restenosis after 10 years.
- Surgical intervention has largely been replaced by percutaneous balloon mitral valvuloplasty. In the relatively few patients who are not suitable, every effort should be made to repair the mitral valve rather than replace it.

Aortic Regurgitation

In aortic regurgitation, there is volume and pressure overload of the left ventricle, eventually leading to contractile dysfunction in the more severe cases. In the chronic situation, many patients remain asymptomatic, despite having moderate or severe regurgitation. Eventually, they become symptomatic with exertional dyspnoea, angina and heart failure.

Key points in diagnosis and management of aortic regurgitation include the following.

- Echocardiography is used to assess LV size and function. The severity of aortic regurgitation is assessed by colour flow mapping of the spatial extent of the regurgitant jet in the left ventricle outflow tract. Patients with mild regurgitation require echocardiographic evaluation every 2 years, whereas those with more severe regurgitation should be studied every 6–12 months.
- Vasodilator therapy can reduce LV dilatation and the regurgitant fraction, slow progression of LV dilatation, and possibly delay the need for surgery. Therapy with nifedipine or ACE inhibitors is recommended for asymptomatic or mildly symptomatic patients with preserved systolic function and moderate or greater degrees of aortic regurgitation.
- Patients with moderate to severe aortic regurgitation who become symptomatic should be referred for surgery. In asymptomatic or mildly symptomatic patients, surgery is indicated if LV function is reduced (LV ejection fraction <55%) or LV end systolic diameter is approaching 55 mm.

- Options for aortic valve surgery are replacement with a mechanical prosthesis, a bioprosthesis, or an aortic homograft. Other options are aortic valve repair and the Ross procedure (pulmonary autograft with homograft replacement of the pulmonary valve).
- Patients who demonstrate good adherence to medications are suitable for replacement with the newer bileaflet mechanical valve prosthesis, which has the best long-term durability and freedom from re-operation. If stable anticoagulation is unlikely to be achieved, an aortic bioprosthesis should be considered. In young female patients a mechanical prosthesis should be avoided, because of the significant risk to mother and foetus posed by anticoagulation during pregnancy.

Aortic Stenosis

Aortic stenosis results from fibrosis and partial fusion of aortic valve cusps, causing progressive obstruction to LV outflow. RHD is an uncommon cause of aortic stenosis and almost always occurs in the presence of associated rheumatic mitral valve disease. The classic symptoms are dyspnoea on exertion, angina, and syncope. Symptoms are gradual in onset, but are usually slowly progressive over time, especially if there is associated mitral valve disease.

Key points in diagnosis and management of aortic stenosis include the following.

- Two-dimensional echocardiography shows the thickened and restricted aortic valve leaflets and allows assessment of LV size and systolic function. Continuous wave Doppler echocardiography is used to calculate the gradient across the aortic valve and the aortic valve area.
- Patients usually do not develop symptoms of exertional dyspnoea and fatigue until a moderate or severe systolic gradient develops (>40–50 mmHg). Once symptoms develop, prognosis is poor without surgery.
- Percutaneous aortic valvuloplasty is reserved only for patients who are not candidates for surgery, as it has a high recurrence rate.
- Aortic valve replacement with a mechanical valve, a bioprosthetic valve, or a homograft is the definitive therapy for symptomatic aortic stenosis. It should be performed in all patients with significant gradients and a reduced valve area once they develop exertional symptoms.

Pregnancy and Rheumatic Heart Disease

Normal pregnancy will worsen the effects of any pre-existing valvular disease. Predictors of increased maternal and foetal risk are reduced LV systolic function, significant aortic or mitral stenosis, moderate or severe pulmonary hypertension, a history of heart failure, and symptomatic valvular disease before pregnancy.

Ideally, patients with known rheumatic valvular disease should be properly assessed before pregnancy occurs. If they are already symptomatic due to significant RHD, serious consideration should be given to intervention prior to pregnancy. In patients with moderate or severe mitral stenosis, percutaneous balloon mitral valvuloplasty should be considered, because of the high risk of maternal and foetal complications during pregnancy. Patients with mechanical valves who are on warfarin should be given appropriate contraceptive advice and should be counselled about the risks to mother and foetus with pregnancy.

Warfarin crosses the placenta but heparin does not. However, there is an increased risk of prosthetic thrombosis with heparin and a risk of embryopathy with warfarin, especially in the first trimester. The choices for antithrombotic therapy during pregnancy are low molecular weight heparin throughout, warfarin throughout, or low molecular weight heparin for the first trimester and then warfarin.

Warfarin throughout pregnancy is the favoured regimen if the dose can be kept to ≤5 mg.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type evidence supporting the recommendations is specifically stated for selected recommendations in the original guideline document.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Accurate diagnosis and appropriate management of acute rheumatic fever and rheumatic heart disease

POTENTIAL HARMS

- Toxic effects of salicylates (tinnitus, headache, hyperpnoea) are likely above 20 mg/100 dL, but often resolve after a few days. There is also the risk of Reye's syndrome developing in children receiving salicylates, who develop certain viral infections, particularly influenza.
- The potential major adverse effects of short courses of glucocorticoids, including gastrointestinal bleeding and worsening of heart failure as a result of fluid retention, should be considered before they are used.
- There is a rare risk of serious allergic reactions to penicillin and fatality as a result of anaphylaxis.
- Intramuscular bleeding from benzathine penicillin G (BPG) injections, used in conjunction with anticoagulation therapy in Australia, is rare.
- The most serious complication of percutaneous balloon mitral valvuloplasty is tearing of the mitral valve leaflets and/or subvalvular apparatus, causing severe mitral regurgitation.
- The main disadvantages of bioprostheses are their limited durability in younger patients (15 to 50 years).
- The main complications of mechanical valves are bleeding and thromboembolic events, usually due to problems with anticoagulation adherence. As with all prostheses, other complications such as endocarditis, prosthetic valve thrombosis, valve dehiscence, and haemolysis may occur.

- Aortic valvuloplasty has significant morbidity and occasional mortality, particularly in elderly patients. Follow-up studies have shown that initial improvement is usually not maintained after a few months. There is a high restenosis rate, particularly in very deformed valves.
- Warfarin crosses the placenta but heparin does not. However, there is an increased risk of prosthetic thrombosis with heparin and a risk of embryopathy with warfarin, especially in the first trimester.

CONTRAINDICATIONS

CONTRAINDICATIONS

- A large left atrial thrombus is a contraindication to percutaneous balloon mitral valvuloplasty (PBMV).
- Angiotensin receptor antagonists and angiotensin-converting enzyme (ACE) inhibitors are contraindicated during pregnancy.

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- This document has been produced by the National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand for the information of health professionals. The statements and recommendations it contains are, unless labelled as "expert opinion," based on independent review of the available evidence. Interpretation of this document by those without appropriate health training is not recommended, other than at the request of, or in consultation with, a relevant health professional.
- The review does not address primary prevention of acute rheumatic fever (ARF) — this area is controversial and the literature is extensive. The authors consider that such discussion would detract from the focus on best practice in the diagnosis and management of ARF and rheumatic heart disease (RHD). Moreover, while there is good evidence for the efficacy and cost-effectiveness of secondary prevention of ARF, there is no clear evidence that systematic, population-wide, sore-throat treatment programs are cost-effective.
- This review provides a general framework and should not over-ride good clinical judgement. Treatment should take into account the patient's comorbidities, drug tolerance, lifestyle, living circumstances, cultural sensibilities, and wishes. When prescribing medication, clinicians should observe usual contraindications, be mindful of potential adverse drug interactions and allergies, and monitor responses and review patients regularly.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Quick Reference Guides/Physician Guides

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness
Staying Healthy

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

RF/RHD Guideline Development Working Group of the National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand. Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia: an evidence-based review. Sydney (Australia): National Heart Foundation of Australia, Cardiac Society of Australia and New Zealand; 2006 Jun. 84 p. [261 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2006 Jun

GUIDELINE DEVELOPER(S)

Cardiac Society of Australia and New Zealand - Disease Specific Society
National Heart Foundation of Australia - Disease Specific Society

SOURCE(S) OF FUNDING

National Heart Foundation of Australia
Cardiac Society of Australia and New Zealand

GUIDELINE COMMITTEE

Writing Group

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Lead Authors: Professor Jonathan Carapetis (*Chair*); Dr Alex Brown; Dr Warren Walsh

Writing Group Members: Dr Keith Edwards; Dr Clive Hadfield; Professor Diana Lennon; Ms Lynette Purton; Dr Gavin Wheaton; Dr Nigel Wilson

Secretariat support: Mr Traven Lea; Ms Kelley O'Donohue

Other reviewers and contributors: Dr Leslie E Bolitho; Dr Andrew Boyden; Dr Christian Brizard; Dr Richard Chard; Ms Eleanor Clune; Dr Sophie Couzos; Dr Arthur Coverdale; Professor Bart Currie; Dr James Edward; Dr Tom Gentles; Professor Marcia George; Dr Jeffery Hanna; Dr Noel Hayman; Dr Ana Herceg; Dr Marcus Ilton; Dr Jennifer Johns; Dr John Knight; Dr John McBride; Dr Malcolm McDonald; Dr Johan Morreau; Dr Michael Nicholson; Dr Ross Nicholson; Ms Sara Noonan; Dr Briar Peat; Dr Peter Pohlner; Dr Jim Ramsey; Dr Jenny Reath; Ms Emma Rooney; Dr Warren Smith; Dr Andrew Tonkin; Dr Lesley Voss; Dr Mark Wenitong; Mr Chris Wilson; Dr Elizabeth Wilson; Dr Keith Woollard

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

ENDORSER(S)

Australasian Society for Infectious Diseases - Disease Specific Society
Australasian Society of Cardiac and Thoracic Surgeons - Professional Association
Australian College of Rural and Remote Medicine - Professional Association
Australian Indigenous Doctors Association - Professional Association
Council of Remote Area Nurses of Australia Inc. - Professional Association
Internal Medicine Society of Australia and New Zealand - Medical Specialty Society
National Aboriginal Community Controlled Health Organisation - National Government Agency [Non-U.S.]
Royal Australasian College of Physicians - Professional Association
Royal Australian College of General Practitioners - Professional Association
Royal College of Nursing Australia - Professional Association

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the National Heart Foundation of Australia Web site.

Print copies: Available from the National Heart Foundation of Australia's national telephone information service at 1300 36 27 87 or E-mail: heartline@heartfoundation.com.au.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- Diagnosis of acute rheumatic fever. Quick reference guide for health professionals. 2006. 4 p. Electronic copies: Available in Portable Document Format (PDF) from the National Heart Foundation of Australia.
- Management of acute rheumatic fever. Quick reference guide for health professionals. 2006. 4 p. Electronic copies: Available in Portable Document Format (PDF) from the National Heart Foundation of Australia.
- Secondary prevention of acute rheumatic fever. Quick reference guide for health professionals. 2006. 4 p. Electronic copies: Available in Portable Document Format (PDF) from the National Heart Foundation of Australia.
- Rheumatic heart disease control programs. Quick reference guide for health organisations. 2006. 4 p. Electronic copies: Available in Portable Document Format (PDF) from the National Heart Foundation of Australia.
- Management of rheumatic heart disease. Quick reference guide for health professionals. 2006. 4 p. Electronic copies: Available in Portable Document Format (PDF) from the National Heart Foundation of Australia.

Print copies: Available from the National Heart Foundation of Australia's national telephone information service at 1300 36 27 87 or E-mail: heartline@heartfoundation.com.au.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on April 9, 2007. The information was verified by the guideline developer on June 27, 2007. This summary was updated by ECRI Institute on September 7, 2007 following the revised U.S. Food and Drug Administration (FDA) advisory on Coumadin (warfarin). This summary was updated by ECRI Institute on March 14, 2008 following the updated FDA advisory on heparin sodium injection.

COPYRIGHT STATEMENT

This NGC summary is based on the original guideline.

DISCLAIMER

NGC DISCLAIMER

The National Guideline Clearinghouse™ (NGC) does not develop, produce, approve, or endorse the guidelines represented on this site.

All guidelines summarized by NGC and hosted on our site are produced under the auspices of medical specialty societies, relevant professional associations, public or private organizations, other government agencies, health care organizations or plans, and similar entities.

Guidelines represented on the NGC Web site are submitted by guideline developers, and are screened solely to determine that they meet the NGC Inclusion Criteria which may be found at http://www.guideline.gov/about/inclusion.aspx .

NGC, AHRQ, and its contractor ECRI Institute make no warranties concerning the content or clinical efficacy or effectiveness of the clinical practice guidelines and related materials represented on this site. Moreover, the views and opinions of developers or authors of guidelines represented on this site do not necessarily state or reflect those of NGC, AHRQ, or its contractor ECRI Institute, and inclusion or hosting of guidelines in NGC may not be used for advertising or commercial endorsement purposes.

Readers with questions regarding guideline content are directed to contact the guideline developer.

Date Modified: 9/15/2008